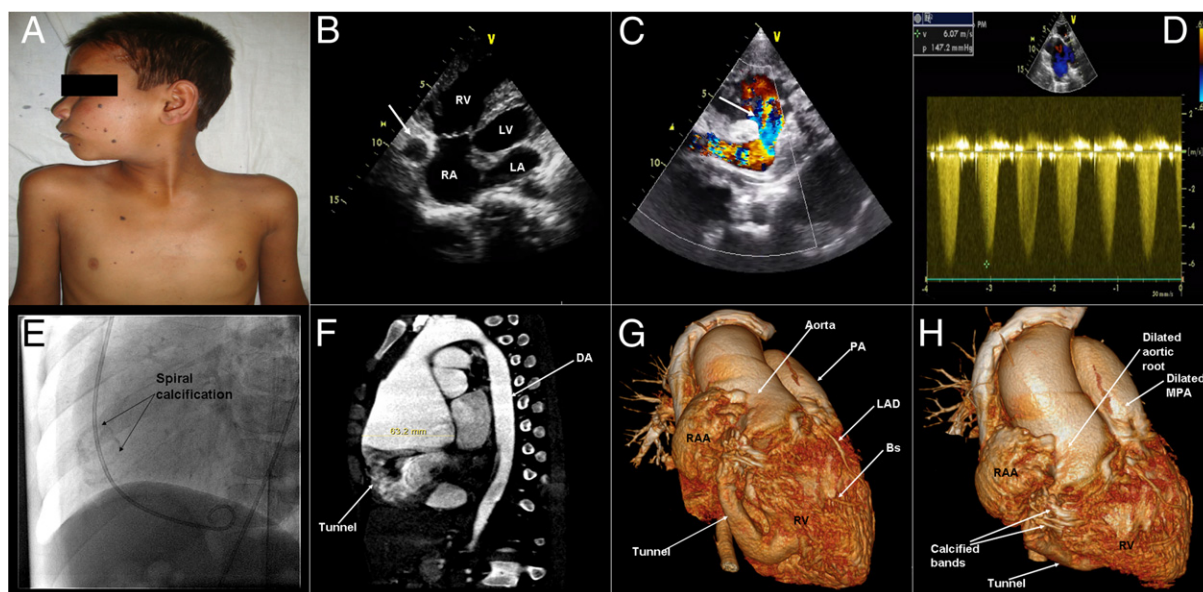


## IMAGES IN CARDIOLOGY

# Calcified Aorto–Right Ventricular Tunnel in a Patient With Multiple Lentigines Syndrome

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**A** 12-year old boy with multiple pigmented nevi and lentigines (**A**) presented to us with symptoms of effort dyspnea and recent onset cyanosis. He had mild thoracolumbar scoliosis, unilateral sensorineural hearing loss, and growth retardation. The 2-dimensional transthoracic echocardiogram revealed a circular echolucency lateral to the tricuspid annulus (**B**, [Online Video 1](#)) (LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle), and color Doppler echocardiogram showed a tunnel-like structure with turbulent jet filling the right ventricle (**C**, [Online Video 2](#)). Two-dimensional echocardiogram with continuous-wave Doppler showed severe valvular pulmonic stenosis (**D**), and contrast echocardiogram revealed right to left shunt across the foramen ovale ([Online Video 3](#)). Cardiac catheterization showed spiral calcification and catheter passage from the aorta to the RV (**E**, [Online Video 4](#)), and the aortogram revealed flow across the tunnel ([Online Video 5](#)). Sixty-four slice computed tomography angiography showed a dilated aortic root (DA) with a tortuous aorto-right ventricular tunnel (**F**). Computed tomography volume-rendered imaging showed a giant tunnel and single left coronary artery giving branches (Bs) to the RV (**G**), with massive dilation of the aortic root and presence of calcified bands at the origin of the tunnel (**H**). LAD = left anterior descending artery; MPA = main pulmonary artery; PA = pulmonary artery; RAA = right atrial appendage.